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Selective photocoagulation in Coats' disease: ten-year follow-up

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PURPOSE. *The diagnostic hallmark of Coats' disease is development of "light bulb" telangiectasis in the retinal periphery, leading to posterior pole intraretinal and subretinal exudation. Even after complete obliteration of all abnormal vessels and resorption of all exudates, follow-up examinations are mandatory for several years.*
METHODS. *We retrospectively analyzed the charts, pictures and/or fluorescein angiographies of a series of 32 consecutive patients, in which the diagnosis of Coats' disease was made. All patients underwent selective photocoagulation of the telangiectasis using a yellow-dye laser. Efficacy of treatment was monitored with drawings and/or fluorescein angiographies.*
RESULTS. *All our patients had unilateral disease, with macular involvement and exudative retinal detachment. Visual acuity improved in one patient, decreased from light perception to blindness in another, and was unchanged in the remainder.*
CONCLUSIONS. *This is the first report of anatomical benefits from treatment with a yellow-dye laser (i.e., selective photocoagulation without cryotherapy or drainage) despite the presence of a severe form of Coats' disease. Our young Coats' patients responded quickly to treatment. With successful obliteration of the abnormal vasculature, exudates began to absorb in about 4 to 8 weeks although, in some cases, 10 to 12 months were needed to obtain resolution of the exudative detachment. Poor visual outcome of 20/100 or worse was common. In Coats' patients, the key issue is continuous control and long-term follow-up. (Eur J Ophthalmol 2002; 12: 501-5)*

KEY WORDS. *Coats' disease, Photocoagulation, Dye yellow laser*

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INTRODUCTION

In 1908, Coats described a condition characterized by retinal vascular changes and exudation (1). Through the years it has frequently been confused with Leber's multiple miliary aneurysms, and in truth probably the two conditions are similar or the same. Clinically, retinal telangiectasia or Coats' disease is usually unilateral and has a marked predilection for young males (2 males to 1 female). The hallmark of this condition is development of "light bulb" telangiectasis in the retinal periphery, leading to posterior pole intraretinal and subretinal exudation. Treatment is directed at obliterating the abnormal telangiectatic vascular changes by photocoagulation or cryotherapy. Because many of those affected are children, treatment usually has to be done under general anesthesia, and cryotherapy is traditionally preferred (2). Cryotherapy is probably superior if large areas of exudates are present beneath the telangiectatic areas, because it is difficult to achieve an adequate photocoagulation reaction when an underlying exudate is present. With the development of the indirect ophthalmoscopic delivery system, however, the role of the laser has be-

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come more relevant because the equipment can now be easily delivered in the operating room, and the treatment given to a child under general anesthesia. Even after complete obliteration of all abnormal vessels and resorption of all exudates, follow-up examinations are mandatory for several years (3,4).

Our study presents the largest series with the longest follow-up of selective anomalous vessel photocoagulation with yellow-dye laser (577 nm) in the presence of exudative retinal detachment.

METHODS

From our Coats' disease database we considered all patients with the presence of a severe form of the disease (i.e., at least one quadrant of exudative retinal detachment with macular involvement), and with a minimum of 10-years follow-up post diagnosis. We excluded patients who could not or would not follow the suggested examination schedule. We report herein a series of 32 consecutive patients who fulfilled our inclusion criteria. Four patients were diagnosed at the San Raffaele Hospital Outpatient Unit, while the remaining patients were each diagnosed at the office of their primary care physician or general ophthalmologist. The diagnosis of Coats' disease was made based on "light bulb" telangiectasis in the retinal periphery with accompanying intraretinal and/or subretinal exudation. All patients were treated and followed at the San Raffaele Hospital by two of the authors (P.N. and F.B.).

We retrospectively analyzed the charts, pictures (drawings and color photographs) and/or fluorescein angiographies of the 32 patients. All patients underwent selective photocoagulation (577 nm) of the telangiectasis using a yellow-dye laser (Coherent, Palo Alto, CA, USA), which has a better hemoglobin absorption characteristic than other dye lasers. The Goldmann lens associated with a 500 µm spot size was changed in the last 9 patients to the panfunduscopic lens. To maintain the 500 µm spot size of the Goldmann lens, the Coherent photocoagulation spot size was adjusted to 200 µm to accommodate the panfunduscopic lens. In all cases, we used variable duration and power, long and high enough to cause closure and/or significant shrinkage of the anomalous vessels.

During laser treatment, patients were propped-up into a seated position on the nurse's lap, while she braced their head on the headrest of the laser machine. An intravenous injection of ketamine hydrochloride was usually sufficient to sedate the child and perform an adequate treatment. In two selected cases, we used endotracheal intubation. Our laser facility is supplied with anesthesiology equipment.

In all patients, the first treatment session was started in 1 to 3 weeks following the diagnosis of Coats' disease. After the laser treatment, dexamethasone eye drops were used four-times daily for a period of 7 - 10 days, to reduce vitreal inflammation. Ibuprofen was prescribed to relieve pain the day of treatment.

Efficacy of treatment was monitored with drawings and/or fluorescein angiography which was performed initially 8 weeks after the treatment, then each four months for the first two years, each year for the next five years, once between the eighth and ninth years, and the last time at ten years (i.e., the close of the study). These follow-up examinations helped to determine whether there was complete and permanent interruption of blood flow, as demonstrated by a marked reduction in the number of exudates. Treatment was considered successful if there was complete occlusion of the feeding arterioles with absorption of the exudates. Retreatment was often necessary to achieve complete closure.

RESULTS

Patient demographics and treatment details during the 10-years follow-up are reported in Table 1. The mean age of the 32 study patients was 4.2 years (range 3-7 years). The distribution was 21 males to 11 females. No patient had hemorrhages during or after treatment. Two cases underwent computerized tomography scans elsewhere for suspected retinoblastoma (patients 10, 16).

Our briefest and longest intervals between consecutive treatments were 45 days and 5 months, respectively. The mean interval between the last laser treatment and complete regression was 13.7 months.

Visual acuity of one patient (patient 14; shown in Fig. 1) improved (mild macular involvement) during the 10-year follow-up; three patients (patients 8, 12 and 19) had 20/80; one (patient 10) had count fingers (CF); one (patient 17) had hand motion (HM); and one

eye with light perception at initial examination developed neovascular glaucoma and phthisis bulbi and had to be enucleated. The remaining 25 patients had a visual acuity of 20/100 or worse.

No association with other systemic and ocular diseases (e.g., muscular dystrophy (5), branch retinal vein obstruction (6), retinitis pigmentosa (7), and retinopathy of prematurity (8)) was detected in our patients. No patient had a family history of Coats' disease.

DISCUSSION

To the best of our knowledge, this is the first report of anatomical benefits of treatment with yellow-dye laser, selective photocoagulation (no cryotherapy or drainage), despite the presence of a severe form (i.e., at least one quadrant of exudative retinal detachment with macular involvement) of Coats' disease as shown in Figure 2, Panels A and B.

TABLE I - PATIENT DEMOGRAPHICS AND TREATMENT DETAILS DURING THE 10-YEAR FOLLOW-UP

Patient number	Age (yr) at first treatment	Total number of treatment spots	Total number of laser treatments	Briefest interval between consecutive treatments	Longest interval between consecutive treatments	Complete regression, interval from last treatment	Final visual acuity
1	3	290	2	3 mos	-	5 mos	20/100
2	7	580	5	45 days	45 days	enucleation	NA
3	4	340	3	2 mos	3 mos	12 mos	20/100
4	5	400	4	45 days	3 mos	18 mos	20/200
5	4	320	3	45 days	3 mos	12 mos	20/200
6	5	380	3	45 days	3 mos	18 mos	20/200
7	4	280	2	2 mos	-	12 mos	20/100
8	3	190	1	-	-	3 mos	20/80
9	4	270	2	2 mos	-	3 mos	20/200
10	6	445	4	45 days	5 mos	2 yrs	CF
11	5	640	6	45 days	3 mos	3 yrs	20/200
12	3	180	1	-	-	3 mos	20/80
13	4	260	2	2 mos	-	5 mos	20/100
14	4	180	1	-	-	3 mos	20/30
15	4	400	3	45 days	3 mos	12 mos	20/100
16	4	260	2	3 mos	-	5 mos	20/200
17	6	720	5	45 days	3 mos	3 yrs	HM
18	4	180	1	-	-	3 mos	20/100
19	3	210	2	45 days	45 days	5 mos	20/80
20	4	320	2	3 mos	-	5 mos	20/200
21	4	445	4	45 days	3 mos	18 mos	20/200
22	6	600	5	45 days	3 mos	3 yrs	20/200
23	4	300	2	3 mos	-	18 mos	20/100
24	3	145	1	-	-	3 mos	20/100
25	4	360	3	45 days	3 mos	18 mos	20/100
26	4	350	3	45 days	3 mos	12 mos	20/200
27	5	350	4	45 days	3 mos	18 mos	20/200
28	3	380	4	45 days	3 mos	18 mos	20/100
29	4	560	4	45 days	3 mos	18 mos	20/200
30	3	250	3	45 days	3 mos	12 mos	20/100
31	3	240	2	45 days	3 mos	12 mos	20/100
32	5	500	4	45 days	3 mos	18 mos	20/200

NA = not applicable; CF = count fingers; HM = hand motion

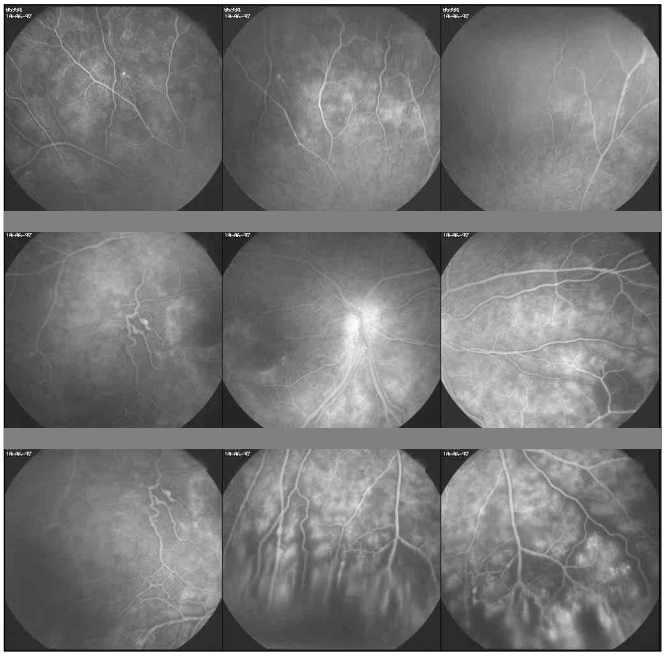

Fig. 1 - *Fluorescein angiographic map of patient 14, a 4-year-old boy with a dilated retinal capillary bed, enlarged intercapillary spaces and retinal capillary nonperfusion in the inferior midperiphery. Retinal exudative detachment was present in the extreme inferonasal periphery, but is not evident in the photos. Mild macular involvement is also evident. This is the only case in which the acuity reached 20/30 after treatment.*

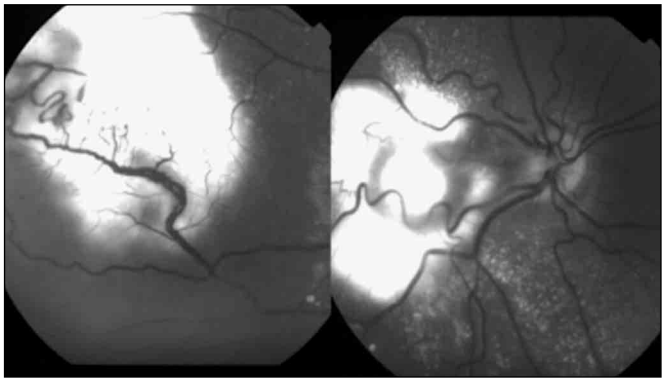

Fig. 2 - *Macular involvement and midperipheral fundus telangiectatic retinal vessel abnormalities can be seen, with hard exudate and serous retinal detachment limited to one quadrant.*

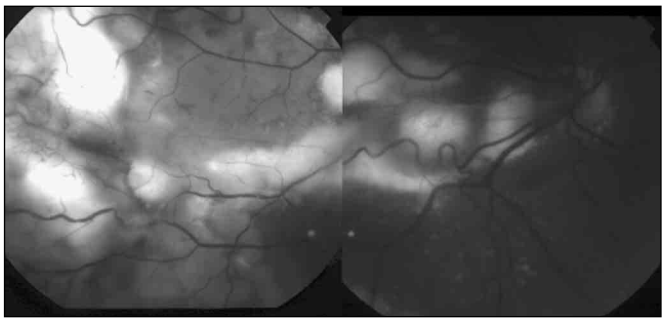

Fig. 3 - *Same fundus as shown in Figure 2 at one month after direct treatment to many of the larger vascular abnormalities. The midperipheral hard exudate has diminished as well as the macular exudate.*

Despite the fact that several manufacturers have developed an indirect ophthalmoscopic delivery system that facilitates peripheral retinal photocoagulation, thus allowing for treatment of patients who previously could not be treated with a slit-lamp system (e.g., pediatric or mentally retarded patients), the slit-lamp system remains our delivery method-of-choice for most cases with extensive vessel involvement, especially those requiring precise laser burn or prolonged exposure to laser light.

Fluorescein angiography is not the most practical way to follow-up these patients. Often, frequent ophthalmoscopic examinations are the best way. With the appearance of new exudate, it can be assumed that new vascular abnormalities have developed. We have seen, as have others (2), recurrences of the disease as long as 30 months after an apparent cure. Such an extended duration between consecutive treatments was not observed among our 32 patients (Tab. I).

Multiple treatment sessions are often necessary. Our briefest interval between consecutive treatments was 45 days. Thus, we are not able to confirm if it is better to wait for at least 3 months before considering additional laser photocoagulation, as Shields et al have suggested in their work (9).

With the successful obliteration of the abnormal vasculature, the exudate will begin to absorb in about 4 to 8 weeks (Fig. 3). As long as 10 to 12 months may elapse before resolution of the exudative retinal detachment. However, frequent follow-up examinations are mandatory.

Patient compliance is very important to treatment success. Parents often believe that doctors over-examine and over-treat the child, and consequently they are prone to skip frequent visits. According to Shields et al (9), the key issue is continuous control and long-term follow-up.

The natural progression in advanced Coats' disease

is toward the development of a blind, painful eye. Spontaneous regression does rarely occur, while some eyes quietly progress to a phthisical state (10-12). In our series, we observed only one eye evolve toward neovascular glaucoma and phthisis bulbi (patient 2).

The prognosis following treatment is best when only one or two quadrants are affected, and diminishes significantly when the disease involves more than 180-degrees of the peripheral retina (2). Visual outcome in patients with thick and dense foveal exudation, is poor. In young adults and older children Coats' disease is less aggressive, and there is less likelihood of progressive exudation and retinal detachment (9). We found that our young Coats' patients responded quickly to yellow-dye laser treatment.

CONCLUSIONS

To the best of our knowledge, this is the first report of anatomical benefit despite the presence of severe Coats' disease, following treatment with selective photocoagulation (no cryotherapy or drainage) using a yellow-dye laser. The slit-lamp remains our delivery method-of-choice for most cases. Our young Coats' patients responded quickly to this treatment, but poor visual outcome of 20/100 or worse is common for this disease. We believe that long-term follow-up will require frequent patient visits to determine if the anatomical benefits of selective photocoagulation with yellow-dye laser, persist or not.

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